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Trends in Hospitalizations for Adults With Congenital Heart Disease in the U.S.

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Objectives The purpose of this study was to better define the epidemiology of hospitalizations for adults with congenital heart disease (ACHD) in the U.S.

Background There is a growing population of ACHD as the result of advances in pediatric care and diagnostic testing.

Methods We used nationally representative data from the 1998 to 2005 Nationwide Inpatient Sample to identify patients ≥18 years of age admitted to an acute care hospital with an International Classification of Diseases-9th Revision code designating a CHD diagnosis. National estimates of hospitalizations and total hospital charges by year were calculated.

Results The number of ACHD hospitalizations increased 101.9% from 35,992 ± 2,645 in 1998 to 72,656 ± 5,258 in 2005. During this period, the annual number of admissions grew for both simple (19,448 ± 1,614 to 44,707 ± 3,644) and complex (12,507 ± 1,172 to 19,973 ± 1,624) diagnoses. The percentage of admissions originating in the emergency department (41.7 ± 0.8%) or involving cardiac surgery (17.7 ± 0.7%) remained stable during the study period. The average patient age (52.3 ± 0.8 years to 53.8 ± 0.6 years, $p < 0.0001$) and proportion of patients with ≥2 medical comorbidities (23.3 ± 0.9% to 33.0 ± 0.7%, $p < 0.0001$) increased. Mean hospital charges per hospitalization increased 127% from $19,186 ± $803 to $43,496 ± $2,166, and the estimated total national charges for these hospitalizations increased 357% from $691 million in 1998 to $3.16 billion in 2005 (in inflation-adjusted 2005 dollars).

Conclusions The number of hospital admissions for ACHD in the U.S. more than doubled between 1998 and 2005. Hospital charges attributable to these admissions have grown even more dramatically. (J Am Coll Cardiol 2009;54: 460–7) © 2009 by the American College of Cardiology Foundation

Congenital heart defects occur in approximately 0.4% to 1% of live births (1–3). These defects range from small septal defects of questionable clinical consequence to often-fatal complex cyanotic lesions (4). Although the incidence of congenital heart disease (CHD) has remained stable during the last 50 years (3), the natural history of most lesions has changed dramatically. The introduction of palliative systemic-to-pulmonary arterial shunts in the 1940s introduced an era of surgical advances which, in concert with improvements in diagnosis and medical care, transformed many of these previously fatal defects into chronic diseases (5–10).

Consequently, there is a growing population of adult patients with CHD (i.e., ACHD). These patients are at variably increased risk for an array of late complications, including heart failure, arrhythmia, and sudden death (11,12). In addition to these specific issues, normal life events such as pregnancy or otherwise-mild illnesses can have exceptional consequences for these patients. It has been estimated that in the year 2000 >400,000 adults in the U.S. were living with a congenital heart diagnosis for which specialized ACHD care is recommended (13).

Hospitalization patterns for adults with CHD in the U.S. have not been thoroughly investigated. The authors of 1 recent study of CHD hospitalizations in the United Kingdom found that although admissions increased across all age groups, this increase was most prominent among adults (14). By using discharge statistics from California, Gurvitz et al. (15) examined hospitalization patterns for patients between 12 and 44 years of age. The transition from pediatric to adult care was associated with decentralization of care, as well as an increasing proportion of admissions via

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the emergency department (ED). Overall national trends in the U.S. remain unknown. In addition, most published studies present data for CHD admissions as a whole but do not provide estimates for specific types of CHD. Given the heterogeneity of CHD diagnoses, these distinctions are important.

As the life expectancy for adults with CHD increases and diagnostic and therapeutic options continue to evolve, significant public and private resources will be required to care for this patient population. Although no national estimate of hospital charges is available, studies have examined factors associated with resource utilization, as estimated by hospital charges and total number of in-patient days, for patients with CHD (16,17). However, these estimates were for children in the U.S. and for a population of adults in Quebec.

In this study, we used a nationally representative database of hospital admissions in the U.S. to provide estimates both for the number of ACHD hospitalizations from 1998 to 2005, as well as for total charges associated with those hospitalizations. Trends for simple and complex CHD are presented, as are data for specific diagnoses.

Subjects and Methods

Data source. We analyzed data from the 1998 to 2005 Nationwide Inpatient Sample (NIS), a subset of the Healthcare Cost and Utilization Project sponsored by the Agency for Healthcare Research and Quality. The NIS is the largest publicly available all-payer inpatient care database in the U.S., including data on approximately 7 to 8 million discharges per year, and is a stratified sample designed to approximate a 20% sample of U.S. community (nonfederal, short-term, general, and specialty) hospitals. National estimates are produced by the use of sampling weights provided. Healthcare Cost and Utilization Project data have been used previously to study trends in hospitalization and resource utilization for CHD in children, as well as for specific ACHD procedures (16,18). The institutional review board of the Hospital of the University of Pennsylvania granted an exemption as this study used anonymous data.

Study population. Our study cohort included adults 18 years of age or older admitted to an acute care hospital for any reason with an International Classification of Diseases-9th Revision (ICD-9) diagnosis code designating a congenital cardiac defect. The ICD-9 codes used include all ICD-9 codes included in Clinical Classifications Software Code 213 (cardiac and circulatory congenital anomalies) except 747.5 (absence of the umbilical artery), 747.6 (peripheral vascular anomalies), and 747.8 (cerebrovascular anomalies).

Study variables. Demographic covariates included age in years, gender, and year of admission. Each diagnosis code was categorized as simple, complex, or unclassified based upon the 32nd Bethesda Conference report (13), as shown in the Online Appendix. Diagnoses classified as moderately or severely complex in the 32nd Bethesda Conference document, for which specialized ACHD care is recommended, are defined as complex in this analysis. The ICD-9 codes representing diagnoses not specifically addressed by that report were categorized, to the extent possible, to be consistent with previously published literature (17). Patients with isolated simple defects and coexisting pulmonary hypertension were categorized as complex.

Abbreviations and Acronyms

ACHD = adult congenital heart disease
ASD = atrial septal defect
ED = emergency department
ICD-9 = International Classification of Diseases-9th Revision
LOS = length of stay
NIS = Nationwide Inpatient Sample
PFO = patent foramen ovale

We defined comorbidities based on Elixhauser's comprehensive set of comorbidities (19). Cardiac comorbidities were excluded, as described by Elixhauser et al. (19), because they may alternatively represent procedural complications. For simplicity, presented results use a comorbidity index (absolute number of comorbidities: 0, 1, and ≥2).

Non-CHD diagnoses were defined by the following ICD-9 diagnosis codes: bacterial endocarditis (421); coronary artery disease (410, 411, 413, 414); electrophysiology diagnoses (426, 427, 785.1); heart failure (428); ischemic stroke or transient ischemic attack (433, 434, 435, 438); pulmonary hypertension (416); and pregnancy (630 to 677). Procedures were defined by the following ICD-9 procedure codes: cardiac surgery with bypass (96.1 to 96.6); implantable cardioverter-defibrillator insertion or revision (379.4, 379.5, 379.6, 379.7); pacemaker insertion or revision (377, 378); atrial septal defect/ percutaneous patent foramen ovale (ASD/PFO) closure (35.52); and percutaneous coronary intervention (00.66, 36.01, 36.02, 36.05, 36.06, 36.07). The primary non-CHD diagnosis was defined as either the primary diagnostic code or the second diagnostic code if the first coded diagnosis designated CHD.

Primary outcomes of interest were the annual number of admissions for all CHD diagnoses as well as annual admissions for subgroups of the overall population such as simple and complex CHD diagnoses. Secondary outcomes of interest were length of stay (LOS) in days and total hospital charges in dollars. The LOS and total hospital charges are reported in the NIS database.

We present absolute numbers of estimated admissions instead of population-adjusted rates for several reasons. First, the U.S. adult population grew relatively little, <10%, between 1998 and 2005. Second, an incidence rate should include in the denominator the number of people at risk for the outcome of interest. Unlike acquired diseases such as heart failure, most adults are not at risk for an admission for CHD, and the number of adults at risk for a CHD admission is unknown. Interpretation of population-based hospitalization rates is further complicated by the potential for multiple admissions for each person in a given year.

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Statistical analysis. Demographic and clinical characteristics were compared by use of the chi-square test for categorical variables and the *t* test for continuous variables. Trends during the study period were calculated with the use of regression models (20). We used linear regression to identify variables predictive of LOS and total hospital charges. Results are presented as estimate ± standard error. All analyses were conducted with SAS version 9.01 (SAS Institute Inc., Cary, North Carolina) statistical software and account for the complex stratified survey design and clustering by hospital (21). Sample weights were used to produce annual national estimates.

Results

There were 35,992 ± 2,645 ACHD admissions in 1998 as compared with 72,656 ± 5,258 in 2005. During the study period, the number of admissions for simple, complex, and unclassified defects increased 130%, 60%, and 98%, respectively. Simple defects increased as a proportion of ACHD admissions from 54.0 ± 1.7% to 61.5 ± 1.4% (p for trend <0.0001), whereas complex diagnoses decreased from 34.8 ± 1.8% to 27.5 ± 1.2% (p for trend <0.0001). There was no change in the percentage of unclassified ACHD admissions, which comprised 11.2 ± 0.6% of admissions in 1998 and 11.0 ± 0.5% in 2005 (p for trend = 0.13) (Fig. 1).

The annual numbers of admissions for individual simple CHD diagnoses are shown in Table 1. Admissions for isolated ASD/PFO increased most steeply, approximately 209% between 1998 and 2005. The proportion of all patients with ASD/PFO who had a coexisting diagnosis of stroke or transient ischemic attack increased steadily from

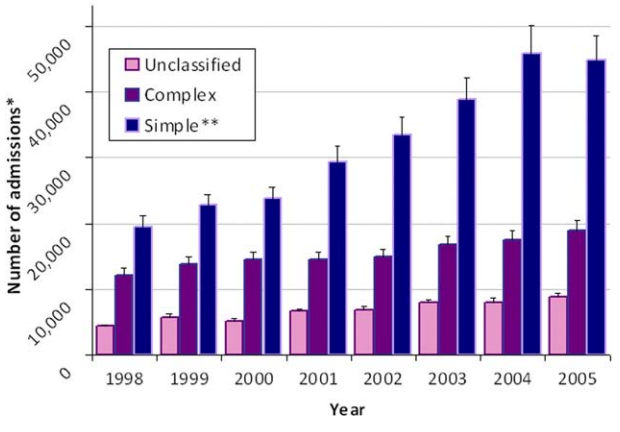

Figure 1 Annual Number of ACHD Admissions in the U.S Categorized by Level of Defect Complexity

*Data represent hospital admissions for adults ≥18 years of age. The analysis used sampling weights to produce nationally representative estimates and accounted for the complex survey design. Error bars represent the standard error. **Classifications are based on the 32nd Bethesda Conference document and other published reports. Simple diagnoses with coexisting complex diagnoses or pulmonary hypertension were classified as complex. ACHD = adults with congenital heart disease.

Table 1 Annual Number of Admissions for Adults With Simple Congenital Heart Disease Diagnoses in the U.S, 1998 to 2005

	1998	1999	2000	2001	2002	2003	2004	2005	Total
Secundum ASD/PFO									
All*	10,916 (936)	12,975 (1,175)	14,063 (1,178)	18,458 (1,680)	22,322 (2,193)	26,140 (2,545)	32,319 (3,548)	30,878 (2,705)	16,8073 (7,992)
Isolated†	8,744 (783)	10,625 (977)	11,557 (989)	15,683 (1,477)	19,184 (1,958)	22,643 (2,270)	28,255 (3,143)	26,979 (2,410)	14,3669 (7,043)
Congenital AI or AS‡									
All	5,285 (617)	6,220 (611)	6,261 (613)	7,624 (742)	7,251 (639)	8,671 (779)	10,101 (1,012)	10,662 (1,093)	62,076 (2,979)
Isolated	4,864 (574)	5,647 (549)	5,712 (564)	7,014 (687)	6,818 (600)	8,002 (721)	9,423 (934)	9,775 (1,005)	57,253 (2,729)
VSD									
All	4,481 (339)	5,055 (381)	5,062 (357)	5,297 (375)	5,515 (385)	5,885 (390)	5,791 (430)	5,932 (406)	43,017 (1,310)
Isolated	3,141 (242)	3,502 (247)	3,417 (245)	3,590 (251)	3,863 (264)	4,064 (263)	4,049 (278)	4,033 (273)	29,658 (822)
Coronary Anomalies									
All	2,476 (265)	2,637 (272)	2,946 (319)	3,114 (319)	3,618 (307)	4,252 (421)	4,184 (358)	4,117 (366)	27,343 (1,296)
Isolated	2,387 (257)	2,516 (259)	2,815 (303)	2,972 (308)	3,474 (300)	4,071 (399)	4,031 (345)	3,968 (351)	26,234 (1,241)
Congenital MR or MS§									
All	821 (110)	902 (110)	546 (66)	697 (76)	628 (144)	566 (79)	524 (66)	741 (173)	5,425 (333)
Isolated	680 (92)	673 (86)	439 (65)	544 (66)	496 (115)	408 (57)	411 (51)	588 (157)	4,240 (272)

Data represent hospital admissions for adults ≥18 years of age. The analysis used sampling weights to produce nationally representative estimates and accounted for the complex survey design. Data are represented as number of admissions (standard error).
*Simple congenital heart disease inclusive of patients with coexisting complex defects or pulmonary hypertension. †Simple congenital heart disease excluding patients with coexisting complex defects or pulmonary hypertension. ‡Includes both congenital AI and AS. §Includes both congenital MR and MS.
AI = aortic insufficiency/bicuspid aortic valve; AS = aortic stenosis; ASD = atrial septal defect; MR = mitral regurgitation; MS = mitral stenosis; PFO = patent foramen ovale; VSD = ventricular septal defect.

23.0 ± 1.4% in 1998 to 31.2 ± 1.3% in 2004 but decreased to 29.5% in 2005 (overall p for trend = 0.0005). The percentage of patients with other CHD diagnoses with a coexisting diagnosis of stroke or transient ischemic attack was significantly lower and remained stable during the study (5.0 ± 0.4% in 1998 to 4.7 ± 0.3% in 2005, p for trend = 0.56).

The annual numbers of admissions for patients with complex CHD diagnoses are presented in Table 2. Tetralogy of Fallot was the most common single complex diagnosis in all years, although the increase in admissions for tetralogy of Fallot was less pronounced than for complex CHD as a whole.

Demographics. Overall, 53.0 ± 0.3% of patients were women, and the mean age was 53.1 ± 0.3 years. The average patient age increased from 52.3 ± 0.8 years to 53.8 ± 0.6 years during the study period (p < 0.0001). In 1998, patients who were ≥55 years of age comprised 43.3 ± 1.6% of admissions. This percentage increased to 49.3 ± 1.2% by 2005 (p for trend <0.0001). Patients who were ≥55 years of age accounted for 50.5 ± 0.5% of admissions for isolated simple defects and 44.0 ± 1.1% of complex defect admissions. Patients with complex defects were younger on average than those with simple defects (51.9 ± 0.5 years vs. 54.8 ± 0.2 years, p < 0.0001) and were more likely to be women (58.9 ± 0.4% vs. 49.9 ± 0.4%, p < 0.0001). Mean patient age increased during the study period for simple (53.5 ± 0.5 years to 55.9 ± 0.4 years, p for trend <0.0001) but not complex (50.3 ± 1.7 years to 51.6 ± 1.2 years, p for trend = 0.20) defects.

Clinical characteristics. Cardiac surgery was performed during 17.7 ± 0.7% of admissions, including 21.6 ± 0.8% of admissions for simple defects and 14.1 ± 0.6% for complex defects. The proportion of ACHD admissions having cardiac surgery remained stable during the study period (18.5 ± 1.3% to 17.2 ± 1.2%, p for trend = 0.08). Cardiac surgery was especially common among patients admitted with a diagnosis of congenital aortic insufficiency or stenosis (42.4 ± 0.1%).

Patients with complex CHD diagnoses had more comorbidities on average than those with simple CHD (1.06 ± 0.02 vs. 1.02 ± 0.01, p = 0.006) and were more likely to have at least 2 comorbidities coded (29.5 ± 0.5% vs. 27.8 ± 0.4%). The number of comorbidities was associated with greater total charges and longer LOS ($29,947 ± $696, $34,556 ± $879, and $37,935 ± $1,106 and 4.3 ± 0.1 days, 5.8 ± 0.1 days, and 7.0 ± 0.1 days for 0, 1, and ≥2 comorbidities, respectively). Patient complexity, as measured by the number of comorbidities, increased between 1998 and 2005. The percentage of patients without any comorbidities decreased from 45.0 ± 1.2% to 35.7 ± 0.8%, whereas those with ≥2 comorbidities increased from 23.3 ± 0.9% to 33.0 ± 0.7% (p for trend <0.0001).

Admissions via the ED constituted 41.7 ± 0.8% of the total, which remained stable during the study period (p for trend = 0.26). Patients having cardiac surgery were much less likely to be admitted through the ED (8.0 ± 0.4% vs. 48.9 ± 0.8%, p < 0.0001). Those patients with simple defects were less likely to present to the ED (40.4 ± 0.9%) compared with unclassified (49.1 ± 0.5%, p < 0.0001) or complex defects (41.4 ± 0.9%, p < 0.0001), although the greater number of admissions for cardiac surgery among patients with simple defects accounted for the difference between simple and complex defects (p = 0.89 after controlling for cardiac surgery).

Trends in specific selected diagnoses and procedures for ACHD admissions are presented in Table 3. In 1998, the primary admission diagnosis was related to CHD or other cardiac disease in 54.4 ± 1.3% of admissions. This proportion had decreased to 50.5 ± 1.2% by 2005 (p for trend = 0.003). Although the number of admissions increased for all the diagnosis groups listed, this increase was most pronounced for arrhythmias (112%) and coronary artery disease (119%). The number of admissions involving pregnancy increased 101%, whereas heart failure, pulmonary hypertension, and bacterial endocarditis admissions increased 83%, 66%, and 51% respectively.

Table 2 Annual Number of Admissions for Adults With Complex CHD Diagnoses in the U.S., 1998 to 2005

	1998	1999	2000	2001	2002	2003	2004	2005	Total
TOF	1,121 (211)	1,082 (137)	1,042 (140)	1,127 (165)	1,110 (135)	1,254 (150)	1,399 (149)	1,248 (155)	9,383 (540)
TGA	603 (223)	842 (159)	611 (95)	839 (170)	703 (162)	831 (142)	801 (139)	1,229 (268)	6,460 (658)
Coarctation of aorta	604 (81)	701 (89)	861 (104)	699 (87)	674 (75)	708 (83)	1,020 (110)	1,128 (145)	6,395 (340)
PS	592 (85)	673 (82)	685 (141)	646 (130)	761 (158)	813 (166)	849 (102)	1058 (114)	6,078 (298)
Combined rare complex*	467 (130)	513 (101)	383 (65)	335 (65)	461 (108)	639 (115)	620 (109)	724 (152)	4,143 (405)
Endocardial cushion defect	442 (77)	562 (92)	488 (74)	455 (73)	581 (80)	513 (78)	554 (74)	545 (92)	4,142 (272)
Ebstein's anomaly	289 (53)	497 (62)	440 (66)	511 (71)	474 (67)	555 (81)	613 (81)	682 (88)	4,062 (241)
Great vein anomaly†	402 (56)	416 (54)	403 (55)	322 (44)	481 (66)	550 (65)	612 (77)	653 (84)	3,839 (213)
Tricuspid atresia or stenosis	391 (163)	408 (74)	323 (67)	407 (66)	380 (100)	412 (85)	420 (87)	527 (116)	3,269 (367)

Data represent hospital admissions for adults ≥18 years of age. The analysis utilized sampling weights to produce nationally representative estimates, and accounted for the complex survey design. Data are represented as number of admissions (standard error). *Composite of admissions data for a group of relatively uncommon complex diagnoses with fewer than 3,000 admissions during the study period, including pulmonary atresia (21.1%), common ventricle (49.4%), hypoplastic left heart syndrome (6.2%), and truncus arteriosus (26.3%). Percentages do not add to 100% because of admissions with multiple diagnoses. †Great vein anomalies include partial anomalous venous return (26.5%), total anomalous venous return (6.2%), and unspecified great vein anomalies (67.3%).

CHD = congenital heart disease; PS = pulmonic stenosis; TGA = transposition of the great arteries; TOF = tetralogy of Fallot.

Table 3 Frequency of Specific Diagnoses and Procedures Associated With Hospitalizations for ACHD

Year	1998	1999	2000	2001	2002	2003	2004	2005	Total
Diagnoses*									
Arrhythmia	11,742 (1,012)	12,717 (1,047)	13,218 (994)	16,332 (1,261)	17,713 (1,352)	20,174 (1,597)	22,458 (1,896)	24,882 (1,961)	139,237 (5,303)
CAD†	8,574 (745)	9,395 (727)	10,770 (853)	12,843 (1,037)	14,275 (1,105)	16,191 (1,279)	18,440 (1,549)	18,788 (1,455)	109,275 (4,235)
Heart failure	7,453 (624)	8,168 (588)	8,389 (584)	9,628 (649)	10,427 (793)	12,1 (908)	12,939 (1,064)	13,604 (1,008)	83,130 (2,905)
Pulmonary hypertension	3,923 (294)	4,747 (402)	4,774 (372)	4,989 (383)	5,617 (446)	6,284 (506)	6,652 (652)	6,528 (525)	43,516 (1,702)
Pregnancy	1,706 (162)	2,217 (204)	2,149 (202)	2,036 (179)	2,568 (226)	2,669 (235)	2,990 (270)	3,431 (312)	19,765 (822)
Bacterial endocarditis	574 (71)	648 (80)	578 (69)	765 (79)	802 (89)	802 (90)	1,103 (116)	867 (99)	6,138 (296)
Procedures									
Percutaneous ASD/PFO closure	134 (90)	165 (59)	308 (133)	627 (150)	2,205 (662)	3,517 (1,008)	4,415 (1,043)	3,219 (566)	14,589 (2,413)
Pacemaker	974 (150)	1,332 (145)	1,310 (148)	1,427 (52)	1,638 (198)	1,798 (191)	2,021 (208)	2,188 (235)	12,687 (674)
PCI	830 (103)	806 (103)	1,065 (136)	1,130 (130)	1,405 (155)	1,832 (198)	1,967 (247)	2,227 (239)	11,262 (633)
ICD	208 (45)	368 (69)	326 (52)	602 (85)	570 (81)	662 (88)	901 (130)	840 (109)	4,476 (312)

Number of patients (standard error). *Data represent national estimates of specific cardiovascular and noncardiovascular diagnoses in hospitalized ACHDs in the U.S. †Data represent national estimates of specific cardiovascular procedures performed in hospitalized ACHDs in the U.S.
CAD = coronary artery disease; ICD = implantable cardioverter-defibrillator; PCI = percutaneous coronary intervention; other abbreviations as in Tables 1 and 2.

The LOS and total hospital charges. The mean LOS was 5.6 ± 0.1 days, with an average total hospital charge of $33,677 ± $803. Although the average LOS remained relatively stable during the study period (5.7 ± 0.1 days to 5.3 ± 0.1 days, p for trend = 0.10), mean hospital charges per admission increased from $19,186 ± $803 to $43,496 ± $2,166. Cardiac surgical admissions incurred greater total charges than medical admissions ($79,500 ± $2,376 vs. $23,892 ± $473, $p < 0.0001$) and were associated with longer LOS (8.5 ± 0.2 days vs. 5.0 ± 0.05 days, $p < 0.0001$). Mean hospital charges per admission increased similarly for both surgical ($44,133 ± $2,480 to $108,195 ± $6,172) and nonsurgical ($13,425 ± $472 to $30,130 ± $1,118) admissions. Admissions for simple diagnoses were shorter but incurred greater charges than those for complex diagnoses (5.6 ± 0.1 days vs. 6.0 ± 0.1 days, $p < 0.0001$; $35,607 ± $884 vs. $33,229 ± $927, $p = 0.0003$). The greater rate of cardiac surgery in patients with simple diagnoses accounted for this latter observation. Adjusting for cardiac surgery, simple diagnoses actually had lower total charges ($p < 0.0001$). Unclassified diagnoses were associated with shorter LOS (4.4 ± 0.1 days) and lower charges ($24,617 ± $798) than simple or complex diagnoses. Total national annual hospital charges for ACHD admissions increased from $691 million to $3.16 billion (all values adjusted for economy-wide inflation to 2005 dollars) (22).

Discussion

The annual number of hospitalizations in the U.S. for adults with CHD more than doubled between 1998 and 2005. Overall hospital admissions in the U.S., by comparison, increased just 13% from 34.7 million in 1997 to 39.2 million in 2005 (23). The disproportionate growth in ACHD admissions is presumably the result of an expanding population of adults with CHD, a demographic trend resulting from several unrelated mechanisms. The first is a paradigm shift during the past 60 years in the surgical and medical approach to children with CHD, which has undoubtedly played the most prominent role in the increase observed in more severe CHD. Improvements in care for adults with CHD, both related to the CHD and comorbidities, have likely also contributed to the population of adults at risk for admission with CHD. The overall rate of mortality for CHD in the U.S. decreased approximately 40% between 1979 and 1997. This decrease was observed for all age groups, although it was most pronounced in those younger than 5 years of age (24). A second reason for the growing number of adults with diagnosed CHD is the availability and use of increasingly sensitive diagnostic tools. Although complex CHD is usually clinically apparent in childhood, subtler and often asymptomatic defects such as small ASD and bicuspid aortic valves easily escaped clinical recognition before advances in noninvasive imaging. Finally, there is increasing awareness of potential relationships between common diseases and congenital heart defects previously

thought to pose little if any risk. The putative relationship between PFO and cryptogenic stroke is the most prominent example.

The size of the population at risk is only one aspect in a complex array determining the number of admissions. An aging population with a greater burden of comorbidities would tend to increase the probability of hospitalization, whereas comprehensive preventive care might attenuate the relationship between population size and the burden of comorbidities with the number of hospitalizations. New therapies may lead to hospitalizations (e.g., percutaneous PFO/ASD closure) or preempt them. Also, events or procedures previously warranting hospitalization are often now managed in the outpatient realm. Further research to define the reasons for the observed increase in admissions is warranted.

Other studies have provided population-based data on adults with CHD in regions of Europe and Canada. Billett et al. (14) evaluated trends in hospitalization for CHD in the United Kingdom. They reported a 16% increase in hospitalizations for a primary diagnosis of CHD across all age groups between 1995 and 2004. Hospitalization rates among adults grew more rapidly. The fastest growth, around 50%, was for 45- to 64-year old patients. Marelli et al. (25) described the prevalence of CHD in Quebec between 1985 and 2000 by using ICD-9 codes. They reported a CHD prevalence of 4.09 per 1,000 adults in 2000, an increase of 14% from 1985. Although the prevalence of severe CHD in adults was much lower, it increased 85% during the study period. By the year 2000, there were more adults than children living with CHD in Quebec.

Patients in this study were older than reported in other epidemiologic studies of ACHD (25). The heterogeneity may in part relate to methodological differences, such as inclusion of relatively mild defects, but we believe the most likely explanation is the population studied. Although many previous studies provided data on the general population of ACHD patients, we focused on hospitalizations. It stands to reason that hospitalized ACHD patients would be older, on average, than the population as a whole.

Our findings parallel the reported dramatic increase in demand for outpatient ACHD care. Gatzoulis et al. (26) described a 269% increase in patient workload between 1992 and 1997 for one specialized outpatient ACHD clinic. Presumably, closer follow-up and appropriate preventive care could attenuate rising emergency department use and hospital admissions for adults with CHD related to a growing population at risk.

The greatest increase in admissions was found for atrial septal defects, an ICD-9 code that includes both true secundum ASD and PFO. Admissions for isolated ASD/PFO more than tripled over the 8 years studied. We suspect that most of this growth reflects the increasing diagnosis of PFO and small ASD. First, PFOs greatly outnumber frank ASD. Although the authors of one study estimated that approximately 188,000 adults in the U.S. had moderate-to-large secundum ASD (two-thirds with previous surgical repair) in 2002 (27), autopsy and echocardiographic studies have reported a PFO prevalence upwards of 20% (28,29). Increased clinical suspicion for PFO in patients with cryptogenic stroke, migraine, and other reportedly related diseases has probably played a role. This finding is supported by the increasing proportion of ASD/PFO associated with a diagnosis of stroke or transient ischemic attack. There was a decrease in the proportion of admissions for PFO/ASD with concomitant complex disease or pulmonary hypertension from 19.9% to 12.6% (Table 1), suggesting an increasing proportion of PFO and hemodynamically insignificant secundum ASD. Admissions for percutaneous ASD/PFO closure are likely partly responsible for the increase (18). The incidental discovery of PFO during imaging for other indications could also increase the likelihood of diagnosis.

The ICD-9 code for congenital aortic insufficiency includes bicuspid aortic valve with or without hemodynamic consequence. Increased use of this diagnosis code for incidentally noted biscuspid aortic valves is unlikely to have had a major impact on the reported findings. First, diagnoses should only be coded if they relate to the hospitalization. The high rate of surgery associated with this diagnosis suggests this was the case. Second, to impact our analysis of trends, there would have to be differential changes in coding between these and other ACHD diagnoses during the study period. Admissions for congenital aortic insufficiency or aortic stenosis paralleled the overall trend, constituting 13.5% of ACHD admissions in both 1998 and 2005.

Adults with complex CHD registered an increase in hospitalizations of approximately 60% between 1998 and 2005. As discussed previously, survival for patients born with complex defects has improved dramatically. One report (25) suggested that the prevalence of complex CHD in adults in Quebec increased almost 85% from 1985 to 2000. Although these patients are fewer in number than those with simple CHD, they are at greater risk for complications related to their underlying defects and surgeries. They are also prone to need further surgical interventions as adults (24). As such, their care often benefits from specialized knowledge and experience (13).

The types of diagnoses and procedures associated with ACHD are shifting. The growing proportion of noncardiac primary diagnoses is notable. A wider array of providers will be exposed to the unique challenges of caring for these patients. Adults with CHD also comprise a growing proportion of those having cardiac procedures, both CHD specific (i.e., ASD/PFO closure) and in general. For example, by using the NIS database, Zhan et al. (30) reported that the number of pacemaker and implantable cardioverter-defibrillator insertions nationwide increased 21% and 136%, respectively, between 1998 and 2004. During the same period, pacemaker and implantable cardioverter-defibrillator procedures for adults with CHD increased 107% and 333%, respectively. Using these data, we estimate that the percentage of implantable cardioverter-defibrillator procedures that

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were performed on adults with CHD increased from 0.7% in 1998 to 1.4% in 2005.

As a result of the increasing number of admissions and increasing charges per admission, ACHD admissions comprise an increasing share of hospital charges. Adjusting for economy-wide inflation, total charges per ACHD admission increased 127% between 1998 and 2005, whereas overall national total hospital charges increased 66.7% (31). After adjusting for economy-wide inflation, national hospital charges for ACHD admissions increased 357% during the study period. Total national hospital charges for all admissions in the U.S., similarly adjusted, increased 82.3% (32). On the basis of these published estimates, ACHD admissions accounted for approximately 0.15% of total national hospital charges in 1998, a number that grew to 0.36% by 2005.

Interestingly, average total charges for each simple or complex CHD admission were comparable. Although adults with simple CHD may be less likely to be admitted to the hospital, the admissions may require comparable resources (17). This finding appears to relate to the reasons for admission, specifically the greater rate of cardiac surgery for patients with simple diagnoses.

Study limitations. The limitations of our data are primarily related to the use of the NIS, a large administrative database, especially its vulnerability to classification error (33). The validity of ICD-9 codes for CHD diagnoses is unknown. By using a similar classification scheme, investigators (17) reported that adults with ICD-9 codes designating complex CHD have greater healthcare resource utilization than those with other diagnoses, which supports the validity of these codes in distinguishing CHD severity on a population level. There is a risk of undercoding and misclassification, given the technical and arcane nomenclature of CHD. As a result, these data likely underestimate the frequency of CHD in hospitalized adults. The ICD-9 codes have limited diagnostic specificity, as witnessed by the lack of distinction between PFO and secundum ASD. Finally, total hospital charges, as reported in the NIS, reflect the amount billed by hospitals. This amount is seldom paid in full by insurers and is not equivalent to costs to the hospital, insurer, or patient. Thus, the figures presented should not be taken to signify the actual societal cost for ACHD admissions. In the absence of significant discrepancies in the ratio of hospital charges to actual costs between ACHD and other types of admissions, however, these data give a sense of the increasing proportion of hospital resources dedicated to patients with ACHD. Although imperfect, total hospital charges should represent a reasonable gross estimate of trends in inpatient resource utilization and has been used by other investigators for this purpose (34). Our data only pertain to hospitalizations, precluding analysis of outpatient services or emergency services not resulting in hospital admission. The striking increase in total hospital charges for adults with CHD constitutes only one facet of this emergent population's economic impact.

Conclusions

These results demonstrate a dramatic increase in hospitalizations for adults with CHD. This growth is likely the result of several unrelated phenomena, most notably improvements in pediatric care and noninvasive diagnostic testing. As a result of the increasing number of admissions and increasing total charges per hospitalization, the care of adults with CHD appears to be claiming an increasing proportion of hospital resources. These changes will affect clinicians, researchers, and health-care policymakers. Health-care providers in all specialties will encounter these complex patients with increasing frequency, and the need for cardiologists with specialized ACHD training will grow. To address the challenges posed by this burgeoning population, we must invest in additional clinical research and health policy planning.

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Key Words: congenital heart disease ■ adults ■ epidemiology ■ hospitalization ■ cardiac surgery.

APPENDIX

For a table outlining the ICD-9 codes used in this analysis, please see the online version of this paper.